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Case report

Pure apraxia of speech due to infarct in premotor cortex

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ABSTRACT

Apraxia of speech (AOS) is now recognized as an articulation disorder distinct from dysarthria and aphasia. Various lesions have been associated with AOS in studies that are limited in precise localization due to variability in size and type of pathology. We present a case of pure AOS in setting of an acute stroke to localize more precisely than ever before the brain area responsible for AOS, dorsal premotor cortex (dPMC). The dPMC is in unique position to plan and coordinate speech production by virtue of its connection with nearby motor cortex harboring corticobulbar tract, supplementary motor area, inferior frontal operculum, and temporo-parietal area via the dorsal stream of dual-stream model of speech processing. The role of dPMC is further supported as part of dorsal stream in the dual-stream model of speech processing as well as controller in the hierarchical state feedback control model.

1. Introduction

Apraxia of speech is a disorder of motor speech planning characterized by slow speech rate, segmentation of syllables, sound distortions, distorted substitutions, trial-and-error articulatory movements, and increased difficulty with increased length and complexity of utterances. It is termed Aphemia in its most severe form. It is distinct from Broca's aphasia given intact repetition, grammar, syntax and writing. It is also distinct from transcortical motor aphasia (TCMA) since articulation is normal on repetition in TCMA. Aphasia may co-occur with aphasia so AOS has been divided into two types – pure AOS or AOS with aphasia. The earlier neuroanatomic correlations in patients with AOS had an infarction in multiple structures around the left inferior frontal gyrus including the adjacent motor cortex, deep white matter, and insula; however these studies had a limited sample size and imaging resolution on CT scans [1]. Further studies on AOS involving a large number of patients and

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improved imaging technique like MRI, emphasized the role of the insular region [2]. However, this ''lesion overlap'' approach was criticized and failed to explain the absence of AOS in all insular strokes. Attempts to explore such neuroanatomical correlation between insula and AOS found no significant association, instead demonstrated either structural damage or hypoperfusion in left posterior inferior frontal gyrus on functional imaging; and few cases had lesions only in precentral and postcentral gyrus [3]. A larger study involving stroke patients with pure AOS demonstrated isolated infarcts in the premotor cortex (PMC) and adjacent motor cortex [4]. This line of evidence reconciled with studies of neurodegenerative AOS where PMC and supplementary motor area (SMA) have been implicated [5]. We present a case of pure AOS with a lesion in dorsal premotor cortex (dPMC) and adjacent motor cortex; and discuss lesion evidence in support of dPMC in speech planning and coordination.

2. Case presentation

A 60-year-old right-handed man with past medical history of ischemic cardiomyopathy with systolic heart failure, hypertension, hyperlipidemia, and gout presented with sudden onset of slurred speech and right arm weakness. Exam showed left upper motor neuron facial palsy and left arm weakness mainly in forearm extensors and wrist extensors. MRI of brain showed an acute infarct in left dorsal premotor cortex and left motor cortex. The infarcts in other areas included the left prefrontal cortex (Fig. 1) and left cerebellum (not shown) were

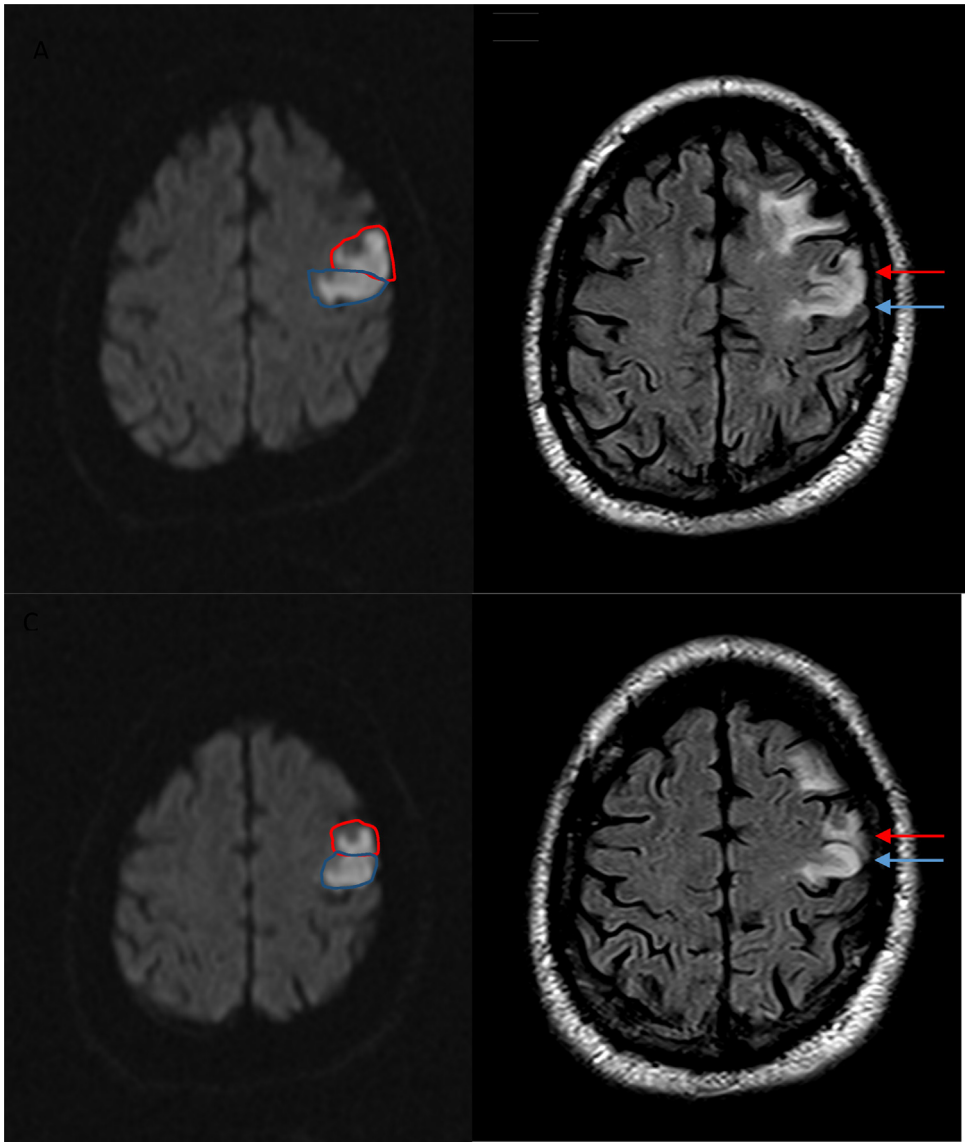

Fig. 1 – MRI Brain. (a) and (c) DWI sequence showing restricted diffusion in left dorsal premotor cortex (anterior outlined region, outlined in red) and left cortical motor cortex (posterior outlined region, outlined in blue). (b) and (d) FLAIR sequence showing infarcts in left prefrontal cortex (arrow head), left dorsal premotor (anterior red arrow) and left motor cortex (posterior blue arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

chronic with no restricted diffusion. The etiology of infarct was likely distal embolization in setting of low ejection fraction due to cardiomyopathy in Rolandic branch of left middle cerebral artery supplying motor cortex and prerolandic branch of left middle cerebral artery supplying premotor cortex.

Upon the initial evaluation, via informal testing the patient followed complex verbal commands normally—speech reception was intact—but he had a significant paucity of speech production, so expressive aphasia was diagnosed by physicians. Often speech disorders are mixed in acute strokes, may co-exist with aphasia before evolving to specific syndrome, or a misdiagnosis of aphasia is made given the overlap of some characteristics with AOS. As spontaneous recovery occurred and with the initiation of speech therapy the true nature of his disorder, apraxia of speech became apparent. The patient was able to repeat spoken phrases though with severely ''garbled'' and difficult to understand speech. Spontaneous, non-stereotyped speech was present and the patient gave appropriate and increasingly lengthy responses to questions though with labored and difficult to understand speech. In addition, through drill type tasks during therapy, confrontational naming, auditory comprehension, repetition, reading and writing was normal without any deficits in grammar or syntax. As noted the patient had no limb apraxia, semantic paraphasias, or pseudobulbar paresis. Further, the patient did not present with buccofacial apraxia. For example, as noted through informal testing to rule out this type of apraxia, the patient was able to correctly mime how to blow out a match, blow a kiss, whistle, smile, and cough. Conversely, and consistent with a diagnosis of AOS, to command the patient could bite his bottom lip, but when asked to produce a syllable or word containing a labiodental phoneme which requires the same movement, he was unsuccessful.

Consistent with a diagnosis of AOS, speech errors included halting output with delayed initiation of phonation along with groping oral movements with silently voiced phonemes. Slow rate of speech, decreased coordination of the respiratory-phonatory subsystems, irregular rhythm and stress, abnormal pauses in words, final consonant deletions, phonemic substitutions, and common articulation errors especially with the fricative phonemes /f/, /v/, /s/, /z/, as well as consonant clusters were all present in his output. His automatic speech (such as counting to 10, stating the days of the week, and singing Happy Birthday) as well as volitional speech were both impaired. For example, in Audio one, recorded on the last day of inpatient rehabilitation (IPR), he was asked to count numbers 1–10. As noted numbers 1–4 were produced without difficulty /wən, tu, θri, fɔr/. The word one begins with a vowel which is easier to produce than a consonant in patients with AOS. The other numbers begin with an initial consonant which are all voiceless even though some are fricative phonemes / θ/ and /f/. Voiceless phonemes are easier to produce than voiced phonemes perhaps due to the longer lag voice onset time in producing voiced phonemes associated with AOS.

As he continued, he begins to make errors. This may be because as the utterance increased the number of errors increased which again is consistent with AOS. Numbers 'five, six, and seven' were hard to say and produced as /su-hai/, /sI/ and /se-en/. It is apparent that he had an anticipatory error with the production of five substituting /f/ for /s/ in preparation for the next two numbers, six and seven. He also consistently omitted the voiced fricative phoneme /v/ in the final position of five and the medial position of seven. He demonstrates a final consonant deletion of /ks/ in the number six which is a combination of a voiceless aspirated stop and voiceless alveolar fricative. Though these phonemes are both voiceless, in producing the number six the patient needed to change lingual positions and move from the front of his mouth for /s/ to the back of his mouth for /k/ and back again to the front for /s/ which is a challenging task for patients with AOS. Numbers nine and ten were produced with deletion of the final consonant /n/ producing /nai/ and /te/. We know that he is able to produce the /n/ phoneme however this patient frequently omitted final consonants. In the repetition tasks of stating ''No ifs ands or buts'', which is a voluntary action, he tries to repeat the phrase but the need to plan and sequence the sounds in order to form each word creates an articulation issue. Motor planning for voluntary speech tasks is more difficult in AOS than involuntary or spontaneous speech. In Audio one he produces /no Its æn bəs/. He utters the word 'no' without difficulty because nasal phonemes are not an issue for him. He substitutes /t/ for /f/ in 'ifs'. The /t/ phoneme is a plosive sound which is considered easier than a more limited closure phoneme such as that involved in the production of the fricative phoneme /f/. One can hear on the Audio his struggle and lag time as he transitions to the next word 'and', he omits the word 'or' completely as well as the phoneme /t/ in 'buts'. In the word 'buts' the final sound is created by a consonant cluster (two or more consonant sounds that come before, after, or between vowel sounds). Consonant clusters are extremely challenging for patients with AOS than producing a single consonant due to the complexity of planning articulatory adjustments between the sounds.

Consistent with a diagnosis of AOS and not expressive aphasia the patient gave a full, rich and elaborate description, as noted on the Audio, of the complex ''Cookie Theft'' picture despite the difficulty in enunciating individual words. However, as can be heard, he states phonetically /si doun wˈʌ fi ou/ / tʃi tʃi geiv əgn gous/, /en Ðʌmʌlɜrs wəʃin dI dIʃIʃ/, /BʌsIn Iz rʌnIn oʊvɜr t frʌn/. In listening to the audio, the patient's first 2 utterances are fairly unintelligible (more frequent sound distortions and omissions than in aphasia) as he attempts to state ''she doesn't want him to fall down. He gave the girl cookies''. However, one can make out the 3rd and 4th production of his description easier, as he was attempting to state ''and the mother is washing the dishes, but the sink is running over the front''. One can hear his struggle to produce the correct sounds within the target words, creating nonsense words and therefore poor effective output. In this segment especially it is clear the patient did not have dysarthria. Patients with dysarthria (a neuromuscular disorder affecting the execution of oromotor and speech functions) consistently produce consonant phonemes imprecisely, whereas with apraxia errors are inconsistent and unpredictable as noted above. When counting he struggled in producing the initial consonant /f/ in five but during his picture description task he produced the /f/ phoneme on more than one occasion. Distortions are the most common type of errors in dysarthria, but in apraxia; repetitions, additions, substitutions, transpositions, prolongations, and omissions are highlighted more

frequently. During the Cookie Theft description, this patient substituted many phonemes, was repetitive on the first word /tʃi tʃi/ in the second phrase, and had a prolongation in the utterance /əgn/ in the second phrase and again frequently omitted the final consonant of many words. One can also determine based on the type of errors this patient produced above, specifically that these were not phonemic paraphasias. In comparison to conduction aphasia (linguistic deficits with phonemic paraphasias and repetition difficulties), patients with AOS produced many errors of transitionalization or blending, while sequencing errors are more typical of the patients with phonemic paraphasia. It has been suggested that phonemic paraphasia represents a breakdown mainly in the retrieval of phonological word patterns, while apraxia of speech is characterized predominantly by a disturbance in encoding phonological patterns into appropriate speech movements [6].

Once he was discharged from IPR, he was then seen for outpatient therapy. Re-evaluation confirmed the diagnosis of AOS and the absence of aphasia, as well as limb or oral apraxia. At this stage, more formal testing (Boston Diagnostic Aphasia Examination (BDAE) short form and Boston Naming Test (BNT)) was completed. Testing indicated moderate-severe apraxia of speech (Table 1).

He was now more aware of his speech errors and often attempted to self-correct. Frustrations were obvious and frequent start–restart behaviors were present. Prosodic deficits, such as stress and intonation, became more apparent as his articulation markedly improved at the sentence level with structured articulation therapy drills and melodic intonation techniques. As noted in the second and third Audio recordings taken during his outpatient therapy course, the patient is describing the same picture at the sentence level in both recordings. The patient states, /wʔʌ t, ɔr, ju:, du:In, sʌn/. He is asking the question ''what are you doing son?'' On his first attempt, he has prolongation of the vowel in the first word 'what' which usually occurs in order to give the motor planning system enough time to organize the next segment of the articulatory program. But the rest of the utterance is fairly clear, yet he has monotonous pitch and limited stress as if he is simply declaring a statement versus a question. During his second attempt, he again states /wʌt, ɔr, dʒu:, du:In, sʌn/. In this attempt he does not have prolongation of the vowel /ʌ/ in the word 'what', perhaps because he has already organized and planned the motor actions of the next segment during his first attempt. He does substitute /ju:/ for /dʒu:/ consistent with irregularities in AOS. But now he inflects the first and final words of the utterance with a slight rise in intonation at the end of the utterance giving more meaning to his listener and creating more of a question versus a statement.

3. Discussion

Our case presented pure AOS associated with acute infarction in relatively restricted area of motor cortex and dPMC. The motor cortex corresponded to the motor homunculus region supplying arm area and explains the pattern of weakness in distal muscles and mostly in extensors. AOS is not a milder form of non-fluent aphasia but emerges as a discrete articulation disorder as the lesion moves away from the inferior frontal gyrus to PMC.

When learning a language, especially the English Language, one has to consider the difficulty in programming voiced and voiceless phonemes. There have been a handful of studies investigating learning English as a second language (ESL) indicating that particular phonemes such as voiceless aspirated stops (a strong burst of breath that accompanies either the release or, in the case of preaspiration, the closure of some obstruents) /p^h/, /t^h/, and /k^h/are found in relatively few languages other than English. These phonemes are not found in languages such as Spanish or French, and therefore will be more difficult for ESL learners to acquire. Eckman suggested native German speakers learning English would have difficulties acquiring voiced contrasts in the word-final position [7]. And in a study by Díaz-Campos, M. investigating Spanish students studying abroad versus those in regular classroom environments in the United States it was concluded that the production of voiced fricatives does not show a pattern of development when learning English as a second language,

Table 1 – Results of speech and language testing.

Informal testing	BDAE short-form	Scores
Speech:	**Comprehension:**	
Non-verbal oral motor tasks with 100%	Word Comprehension:	15/16
Intelligibility: ~50% at sentence level in	Basic Commands:	10/10
unknown contexts	Complex Ideational Material:	5/6 with repetitions
Oral reading sentences with 70% intelligibility		
Sentence production task given picture stimuli with contextual use of pacing strategy to 65% intelligibility		
	Expression:	
	Automatized Sequences:	4/4
	Repetition of Single Words:	3/5; articulatory breakdowns with multi-syllabic words
	Repetition of Sentences:	0/10
	Responsive Naming:	10/10
	(BNT: Short form)	
	Confrontation Naming:	10/15; increasing to 12/15 with phonemic cues; articulatory errors noted

which suggests that fricatives are difficult to acquire. These findings suggest that the motor planning complexity of particular phonemes and producing the precise and timely movements of the articulators for speech prove difficult in non-English speaking persons even without insults to the premotor cortex [8].

The role of PMC in AOS is supported by dual-stream model of speech processing. This model has proposed two streams (i.e. dorsal and ventral) for cortical speech processing to explain observed double dissociations between speech perception and speech recognition [9]. The ventral stream comprises of structures in the posterior middle temporal lobe/temporoparietal area, anterior temporal pole and inferior frontal gyrus; is involved in speech recognition by translating speech signals into semantic representation i.e. ''sound to meaning''. Lesion of the ventral lexicon in temporoparietal area may lead to transcortical sensory aphasia, a syndrome characterized by poor verbal comprehension but preserved repetition and production [10]. Lesion of the ventral lexicon at level of temporal pole may lead to semantic aphasia, most evident in semantic type of fronto-temporal dementia.

The dorsal stream serves sensorimotor integration function in speech perception by translating speech signals from posterior temporal lobe into articulatory representations in the dPMC by maintenance of sublexical representations in an active state during the performance of the task like phonetic discrimination i.e. ''sound to action''. Speech perception tasks involve some degree of executive control and working memory, which might explain the association with frontal lobe. The dorsal stream comprises of dPMC connected to posterior superior temporal lobe via angular gyrus through superior longitudinal fasciculus branch 2 (SLF-II) and superior longitudinal fasciculus branch temporo-parietal part (SLF-tp), traveling above and thus sparing the extreme capsule and insular cortex in left hemisphere. Thus, insula is not part of dorsal stream but there are separate connections between auditory cortex and PMC that travel in extreme capsule underneath the insular cortex. In studies where AOS has been found with insular lesion with or without involvement of inferior frontal gyrus, the deficit might be result of damage to connections of insular region with PMC, rather than a causal involvement. Unlike conduction aphasia, a syndrome that is characterized by good fluency and comprehension but frequent phonemic errors in speech production; AOS is characterized by impaired fluency and perception which is further supported by evidence showing impairment of phonetic discrimination by disruption of PMC by using rTMS, supporting its independent causal role [11].

From an integrated motor control and psycholinguistic approach, a hierarchical state feedback control model has been suggested [12]. The motor control approach consists of a state feedback control where a controller gets combined feedback on state estimation by both the internal feed-forward system that receives an efferent copy from the controller before the execution of action to predict and correct the action and the external feedback from the effector for further correction after the execution of action. In this state feedback control model, hierarchy is introduced from a psycholinguistic perspective at two levels: a lexical – conceptual level and a phonological level. AOS can be explained as dysfunction of controller while in contrast to AOS, dysarthria is a consistent and predictable articulation disorder due to power-issue rather than controller-issue caused by paresis in the effector which is the bulbar musculature supplied by motor fibers including upper motor neurons of corticobulbar fibers originating from the ventral part of the primary motor cortex (spastic dysarthria) or lower motor neuron originating from respective cranial nerve nuclei after termination of corticobulbar tract (flaccid dysarthria) [13]. The hierarchical state feedback control model also better explains the conduction aphasia by reconciling the differences in impaired phonemic planning and intact speech perception by attributing them to separate phonological input and output systems.

In dissecting this patient's speech errors a definitive diagnosis of AOS was determined early on. More specifically [14], kinetic apraxia in which the initiated movement is correct, but the emphasis is on the clunky, not smooth execution of the movement. AOS in which poorly formed phonemes are most salient seems to be like a form of kinetic apraxia. The correct utterance is initiated, the correct phonemes are selected, but the articulatory movements to produce well-formed phonemes, consonant clusters or syllables are not executed correctly. The GMP of an articulation unit itself seems to be damaged. It is important to get the appropriate diagnosis and distinguish between a speech disorder (apraxia or dysarthria) versus a language disorder (such as aphasia) in order to implement more specified therapy techniques [15]. These techniques will then target the presented symptoms previously noted. Speech therapy can be targeted to motor learning and speech perception. By initiating proper therapy, especially in the early stages, it may aid in enhancing the recovery process, achieving a more favorable outcome, and minimizing functional disability. The superficial location dPMC makes it a potential target for non-invasive stimulation in treatment of AOS in the future.

Conflict of interest

None declared.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.pjnns.2017.08.006.

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